Non-Hodgkin's Lymphoma

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Non-Hodgkin's Lymphoma

- Non-Hodgkin's lymphomas (NHL) are a heterogeneous group of malignant lymphomas. There are many different subtypes, every few years the classification is updated. Today, morphology, immunophenotype, molecular, cytogenetics, and other techniques are used for diagnosis.
- Treatment generally depends on the aggressiveness of the disease (indolent, aggressive, or very aggressive)
- Current ICD-9-CM diagnosis code range 200.0_ – 200.8_ and 202.0_ – 202.9_

Behavior

- Indolent – these lymphomas grow slowly. The majority of NHLs are considered indolent. Indolent lymphomas are generally considered incurable with chemotherapy and/or radiation therapy.
- Aggressive – these lymphomas have a rapid growth pattern. This is the second most common form of NHL and are curable with chemotherapy.
- Very Aggressive – these lymphomas grow very rapidly. They account for a small proportion of NHLs and can be treated with chemotherapy. Unless treated rapidly, these lymphomas can be life threatening.

WHO/REAL Classification of Lymphoid Neoplasms

B-Cell Neoplasms

- Precursor B-cell neoplasm
  - Precursor B-lymphoblastic leukemia/lymphoma (precursor B-acute lymphoblastic leukemia)
- Mature (peripheral) B-neoplasms
- B-cell chronic lymphocytic leukemia / small lymphocytic lymphoma
- B-cell prolymphocytic leukemia
- Lymphoplasmacytic lymphoma‡
- Splenic marginal zone B-cell lymphoma (± villous lymphocytes)*
- Hairy cell leukemia
- Plasma cell myeloma/plasmacytoma
- Extranodal marginal zone B-cell lymphoma of MALT type
- Nodal marginal zone B-cell lymphoma (± monocytoid B cells)*
- Follicular lymphoma
- Mantle cell lymphoma
- Diffuse large B-cell lymphoma
  - Mediastinal large B-cell lymphoma
  - Primary effusion lymphoma†
- Burkitt's lymphoma/Burkitt cell leukemia§

T and NK-Cell Neoplasms

- Precursor T-cell neoplasm
  - Precursor T-lymphoblastic leukemia/lymphoma (precursor T-acute lymphoblastic leukemia
- Mature (peripheral) T neoplasms
  - T-cell chronic lymphocytic leukemia / small lymphocytic lymphoma
  - T-cell prolymphocytic leukemia
  - T-cell granular lymphocytic leukemia[II]
  - Aggressive NK leukemia
  - Adult T-cell lymphoma/leukemia (HTLV-1+)
  - Extranodal NK/T-cell lymphoma, nasal type#
  - Enteropathy-like T-cell lymphoma**
  - Hepatosplenic γδ T-cell lymphoma*
  - Subcutaneous panniculitis-like T-cell lymphoma*
  - Mycosis fungoides/Sézary syndrome
  - Anaplastic large cell lymphoma, T/null cell, primary cutaneous type
  - Peripheral T-cell lymphoma, not otherwise characterized
  - Angioimmunoblastic T-cell lymphoma
  - Anaplastic large cell lymphoma, T/null cell, primary systemic type

Hodgkin's Lymphoma (Hodgkin's Disease)

- Nodular lymphocyte predominance Hodgkin's lymphoma
- Classic Hodgkin's lymphoma
  - Nodular sclerosis Hodgkin's lymphoma (grades 1 and 2)
  - Lymphocyte-rich classic Hodgkin's lymphoma
  - Mixed cellularity Hodgkin's lymphoma
  - Lymphocyte depletion Hodgkin's lymphoma

‡ Formerly known as lymphoplasmacytoid lymphoma or immunocytoma

II Entities formally grouped under the heading large granular lymphocyte leukemia of T- and NK-cell types

* Provisional entities in the REAL classification

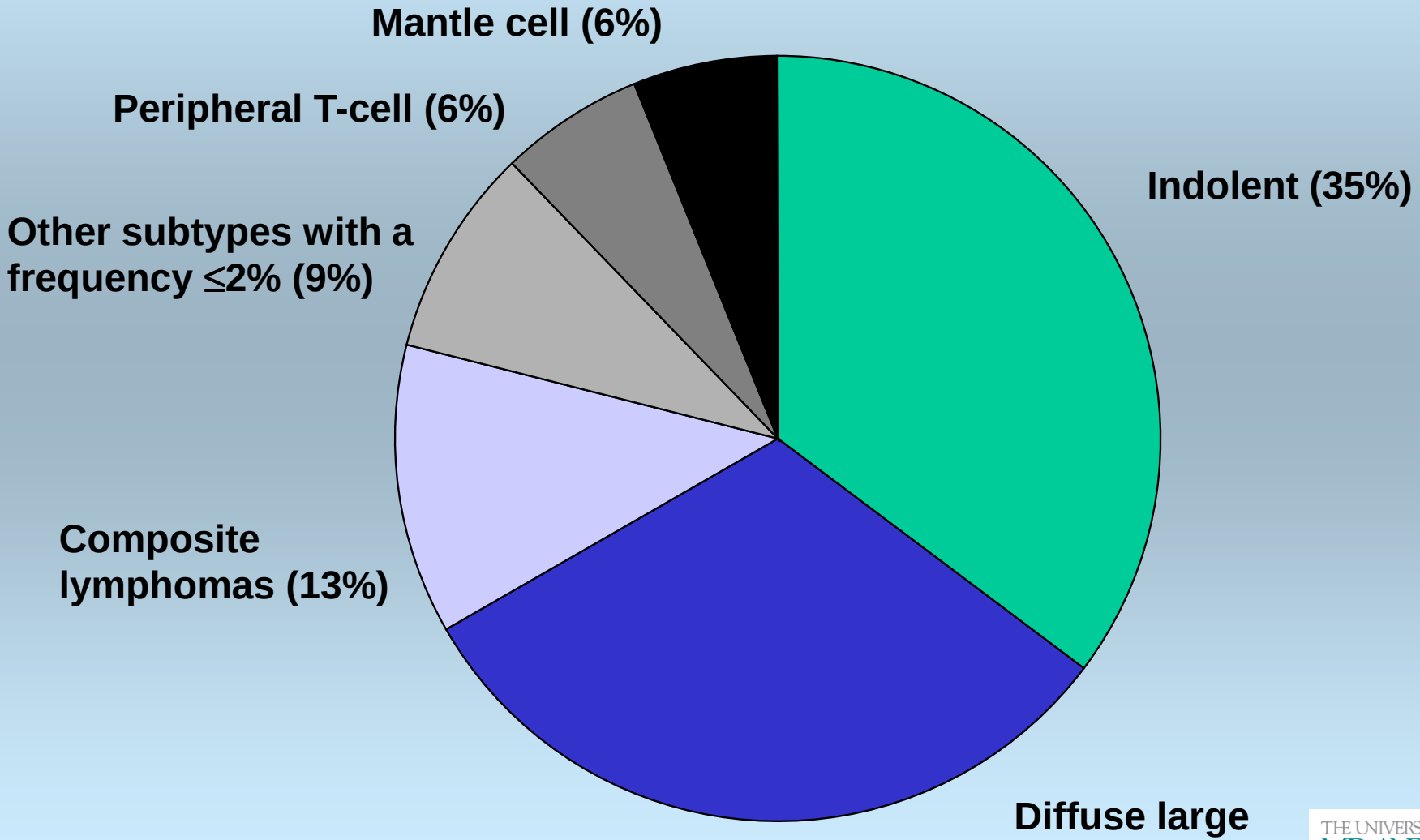
† Not described in REAL classification

§ Includes the so-called Burkitt-like lymphomas

** Formerly known as intestinal T-cell lymphoma

Formerly know as angiocentric lymphoma

Frequency of NHL Subtypes in Adults



Armitage et al. J Clin Oncol. 1998;16:2780–2795.

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Marginal Zone Lymphoma

- Indolent
- Currently codes to 202.8_
- Accounts for ~10% of all lymphomas
- Subcategories
  - MALT (XRT?)
  - Nodal
  - Extra Nodal
  - Splenic

Mantle Cell Lymphoma

- Aggressive
- Currently codes to 202.8_
- Accounts for ~ 6% of all lymphomas
- Considered “incurable” with traditional RX
- Stem cell transplant is offered often as front-line consolidation treatment in “younger” patients

Primary CNS Lymphoma

- Aggressive
- Currently codes to 202.8_
- Accounts for ~ 1-2% of all lymphomas
- Different chemotherapy treatments
- Often requires radiation to the brain:
  - » Brain dysfunction in younger patients
  - » Dementia in older patients

Anaplastic Large Cell Lymphoma (ALCL)

- Aggressive
- Currently codes to 202.8_
- Accounts for ~ 2% of all lymphomas
  - Two groups:
    - ALCL ALK-1+ better prognosis, more common in younger patients and children
    - ALCL ALK-1-negative : as bad as any other T-cell lymphoma

Peripheral T-cell Lymphoma

- Aggressive
- Currently codes to 202.8_
- Accounts for ~ 7% of all lymphomas
- Worse prognosis, often associated with extranodal presentation
- Often requiring salvage treatment and transplant

Large Cell Lymphoma

- Very Aggressive
- Currently codes to 200.0_
- Accounts for ~ 31% of all lymphomas

Other Recommendations

- Changes to terminology
- Changes in disease process

Questions?